correlation with survival (8) It is necessary to define criteria for the best selection and timing for candidates to lung transplantation.
Registries are an important source of data to investigate clinical and research questions. This review points out that only in some instances (incidence/survival analysis) CF Registries are fully able to answer to questions relevant for research. The main limits concern the analysis of historical cohorts in spite of the large number of patients registered. It should be demanded to standardize definitions of variables, outcome measures and timing of data collection to make results comparable among Registries.

P5 USE OF NIV IN THE DRAINING THERAPY: CLINICAL CASE

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Studies show that the use of NIV, in Cystic Fibrosis, has a positive effect on the chronic respiratory insufficiency in patients affected by advanced pulmonary disease and/or in waiting lists for a pulmonary transplant, by improving gas transfers and reducing muscular work.
For this reason we decided to use NIV as support to the draining therapy, focusing on the following aims:

- improving the compliance of the patient;
- improving the efficacy of the physiotherapeutic treatment;
- estimating the course of the illness.

Clinical case: a male patient, 10 years old, affected by Cystic fibrosis with pancreatic insufficiency and chronically colonized by Pseudomonas aeruginosa. The data in the table referto the 12 months before NIV and to the NIV* months.

	before NIV	NIV* months
Physiotherapeutic activity	2–3 sessions per day with PEP-Mask	1 session of NIV and 1 session of PEP-Mask
Height	130 cm (P) → 132 cm (P) +	132 cm (P) → 137.5 cm (P) +
Weight	28 (P) → 29 Kg (P) +	29.2 Kg (P) → 32 Kg (P) +
Spirometry	FEV_1 57.7%	FEV_1 68.4%
Sputum	40 cc with stamps	40cc + fluid without stamps
6 WT	500 mt. with breaks for cough	540 mt. without cough
RX	19	9
Period in bed	52 days	No day
Therapy period	56 days (ev) 94 days (os)	16 days (ev) 25 days (os)
Compliance	VAS 2	VAS 6
Symptomatology	wheezing cough, tiring expectoration, chest tightness	easy expectoration, no chest tightness. Also improved PEP session
Tolerance of effort	Reduced during physical activity	good (gym)

*Resmed pressometric ventilator VPAP III ST-A with nasal mask.

Draining session with 4–5 cycles if C-PAP per 3 days with 7 cm H_2O → use of Coach 2 (respiratory incentive), 3 fast inspirations and 3 slow ones. → Huffing
Conclusions: the use of the ventilator during a draining therapy produced the following results:

- *Improving the compliance*: this was possible because the draining therapy has been less tiring, faster, with no chest tightness, with a better approach of the patient to practise the draining session and to require it in case of obstruction.
- *Obtaining a better efficacy of the physiotherapeutic treatment*: a more flowing sputum, a relevant reduction of the air instability, a better tolerance of effort.
- *Recording positive effects on the course of the illness*: spirometric increase, therapy period reduction, no admission, better saturated-ponderal increase, radiological improvement.

P6 COMPARISON OF HIGH FREQUENCY CHEST WALL OSCILLATION (HFCWO) AND PEP-MASK IN HOSPITALIZED PATIENTS WITH CYSTIC FIBROSIS (CF)

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Background: Reducing bronchial obstruction plays a primary role in the rehabilitation of patients with CF. In the last decades treatment has shifted gradually from passive and tiring techniques to methods that foster self-management of pulmonary disease, including the use of mechanical tools to loosen secretions. However, the greater efficacy of such mechanical methods compared to other procedures has not been demonstrated clearly.
Aim: To compare the efficacy of HFCWO with that of PEP-Mask in the clearance of bronchial tubes from secretion.
Methods: 10 patients with CF (4 males, average age 21.4±7.5 years) hospitalized for a relapse of broncho-pulmonary disease. During hospitalization (average length, 6.1±2.7 days) each patient had daily applications of bronchial disobstruction, one with PEP-Mask and one with HFCWO, each lasting 30 minutes, following standard procedures. The sequence of the two interventions as changed daily. All patients were treated with intravenous antibiotics and aerosol bronchodilators. Oxygen was provided to maintain adequate saturations (SpO2>90%). The following parameters were evaluated: quantity of ejected humid mucus, SpO2, and heart rate (HR) during each application of draining physiotherapy, dyspnea and mood assessment using a visual analogue scale (VAS) at beginning and end of each treatment, FEVI and WT6min at the start and end of hospitalization, and end-study evaluation questionnaire. Statistical significance was evaluated with the Paired T-Test and Wilcoxon Signed Rank Test.
Results: Minimum SpO2 during PEP-Mask was 89.3±3.1 vs. 87.2±3.5 during HFCWO ($p < 0.01$); average quantity of sputum per treatment was 16±9.6 with PEP-Mask vs. 10.3±6.5 ml with HFCWO ($p < 0.02$); HR peak during PEP-Mask was 116.5±8.8 b/m vs. 120±6.7 b/m for HFCWO ($p = 0.08$); evaluation questionnaire showed a perceived greater draining efficacy with PEP-Mask vs. HFCWO ($p < 0.02$).
No significant differences were identified for the other parameters.
Conclusions: Compared to HFCWO, treatment with PEP-Mask is associated with greater draining efficacy, less desaturation, and milder tachycardia in patients with CF. Studies with larger sample size and longer follow up are warranted.

P7 EFFICACY OF HIGH FREQUENCY CHEST COMPRESSION (HFCC) IN THE CLEARANCE OF MUCUS IN PATIENTS WITH CYSTIC FIBROSIS (CF)

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Aims: To determinate the efficacy of HFCC (using the VEST) in the clearance of mucus, by comparing it to the amount of expectorated mucus obtained with PEP-mask. HFCC is made through a system with an inflatable vest, that rapidly inflates and deflates to compress and release the chest wall, creating an airflow inside the lungs. This process moves mucus towards the larger airways, where it can be coughed up and expectorated. PEP-mask is a positive expiratory pressure generated through a face-mask.
Methods: 10 CF patients (M:W=8:2; mean age=24.3 yrs, range=9–43 yrs), clinically stable and undergoing their usual therapies (mean FEV1=54.79%, range=26–93), were randomly recruited. They came twice a week in a 2 weeks period to perform both VEST and PEP-mask therapy each week (1st day VEST, 2nd day PEP, 3rd day PEP, 4th day VEST).
%SaO2 and cardiac frequency were checked with pulseoximeter every 2 min during each session. We evaluated the emotional state and the fatiguing using VAS scales, before and after each treatment. The dry expectoration was weighted in laboratory. HFCC consisted on a VEST treatment of 30 min with 3 phases: the first consisted of 7 min in a low frequency (8 Hz) and pressure (P = 4), the second consisted of 7 min at 10 Hz and P = 5, and the third of 7 min at 15 Hz and P = 6. Every phase was followed by a pause of 3 min where they did 3 cough bouts and expectorated. They did a total of 9 cough bouts followed by expectorations.
PEP-mask consisted of a 30 min treatment. Patients did 7 min of PEP (with a pause of 20 sec every 2 min) followed by a pause of 3 min where they did 3 cough bouts and expectorated for 3 times. They did a total of 9 cough bouts followed by expectorations.
Results: See the table.

	VEST	PEP-mask	P value
Sputum gr	34.09±10.28	34.91±9.84	0.483
% SaO2 mean	95.18±1.79	95.49±1.38	0.293
Peak %SaO2 desat	91.90±3.29	91.30±3.05	0.441
% pz VAS amusing	30	10	
% pz VAS fatiguing	45	45	

Conclusions: VEST is as much effective as PEP-mask technique in clearing sputum from airways of CF patients. No differences neither in the amount of sputum expectorated nor in peak SaO2% desaturation were showed. VEST was perceived as less distressing than PEP-mask, in the same fatiguing condition. So the VEST may be used as substitute or integrative to PEP, to create an alternative to the classic chest physical therapy.
Reference(s)

JC Darbee, JF Kanga, Physiologic evidence for HFCWO and positive expiratory pressure breathing in hospitalized subjects with cystic fibrosis. Physical Therapy, Vol. 85(12), 2005, 1278–1289

P8 AUTOMATION OF CFTR RE-SEQUENCING: TIME AND COSTS ANALYSIS

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Among methods for mutational scan of Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) gene, direct sequencing ensures the highest degree of de-